CHAPTER 65

Coordination and Cerebellar Testing

KEY TEACHING POINTS

- The four cardinal findings of cerebellar disease are ataxia, nystagmus, hypotonia, and dysarthria.
- Testing for cerebellar signs is possible only when motor power is preserved, because the function of the cerebellum is to *administer* movement (i.e., with diminished motor power, there is nothing for the cerebellum to administer).
- There are four cerebellar syndromes, each defined by the patient's bedside findings: cerebellar hemisphere syndrome, anterior cerebellar syndrome, pancerebellar syndrome, and cerebellar infarction. Each syndrome implies a distinct etiology.

I. INTRODUCTION

In the 1920s, after closely observing patients with cerebellar tumors and World War I soldiers with gunshot wounds to the posterior fossa, the British neurologist Gordon Holmes concluded that four physical signs were fundamental to cerebellar disease: ataxia, nystagmus, hypotonia, and dysarthria.[1-5]

II. THE FINDINGS

A. ATAXIA

Ataxia refers to incoordinated voluntary movements that lack the speed, smoothness, and appropriate direction seen in the movements of normal persons. Because the cerebellum's role is to organize and administer movement, testing for ataxia is possible only in patients with adequate motor strength (i.e., 4 or 5 on the MRC scale; see Chapter 61). Tests of ataxia include observation of the patient's gait (see Chapter 7), the finger-nose-finger test, heel-knee-shin test, and rapid alternating movements.

1. FINGER-NOSE-FINGER TEST

In this test, the seated patient takes the index finger of his or her outstretched hand and alternately touches his or her nose and the clinician's index finger being held a couple of feet away. The patient with cerebellar disease may misjudge the range of movement (i.e., **dysmetria**), thus overshooting the target (i.e., **hypermetria**, as in missing the nose and slapping the hand into his or her own face) or undershooting the target (i.e., **hypometria**, as in stopping before reaching the clinician's

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finger). The patient's finger may also deviate from a smooth course, especially if the clinician shifts the target during the test. As the patient's finger approaches the target, an increasing side-to-side tremor may appear (i.e., intention tremor or kinetic tremor). Nonetheless the term *intention tremor* can be confusing because it is applied to two distinct tremors, one of cerebellar disease and another of any action tremor that worsens as the hand approaches a target (e.g., the essential tremor that worsens as a soupspoon or cup approaches the patient's mouth; see Chapter 66). The intention tremor of cerebellar disease, however, is markedly *irregular*, with a large amplitude and low frequency (i.e., less than 5 Hz); it is associated with dysmetria. The intention tremor of essential tremor, by contrast, is *regular*, fine, rapid (8 to 12 Hz), and unassociated with dysmetria.[6]

2. HEEL-KNEE-SHIN TEST

In this test, the supine patient places the heel of one leg on the opposite knee and then slides it down the shin. Like the finger-to-nose test, a positive response may reveal any combination of ataxia, dysmetria, and intention tremor.

Decomposition of movement denotes an abnormal sequence of actions. For example, during the heel-knee-shin test, the patient may completely flex the hip before beginning to bend the knee, thus lifting the heel abnormally high in the air before lowering to complete the movement.[2]

3. RAPID ALTERNATING MOVEMENTS

Difficulty with rapid alternating movements is called dysdiadochokinesia (Babinski coined the original term *adiadochokinesis*).[3] The usual test is rapid pronation and supination of the forearm, but other tasks such as clapping the hands, tapping a table, or stamping the foot are just as good.[3] In all these tests, the movements of patients with cerebellar disease are slower and significantly more irregular in rhythm, range, and accuracy.

B. NYSTAGMUS

1. DEFINITION

Nystagmus is an involuntary to-and-fro oscillation of the eyes. Nystagmus may be congenital or acquired, and the movements may affect both eyes (*bilateral*) or just one eye (*unilateral*). Bilateral nystagmus may be *conjugate*, which means that both eyes have identical movements, or *dissociated*, which implies separate movements. Nystagmus may be *pendular*, which means that the to-and-fro movements have the same velocity, or *rhythmic*, which means that the movement is slow in one direction and quick in the other (rhythmic nystagmus is usually called jerk nystagmus). **Jerk nystagmus** is named after the direction of the quick component (e.g., right conjugate jerk nystagmus). Finally, the direction of the nystagmus may be horizontal, vertical, or rotatory.

2. PATTERNS OF NYSTAGMUS

Although nystagmus is a complicated subject that sometimes defies general principles,* several well-recognized patterns are described below.

A. CEREBELLAR NYSTAGMUS

The most common nystagmus of cerebellar disease is a conjugate horizontal jerk nystagmus on lateral gaze. (See the section titled "Clinical Significance".)

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*One famous neuro-ophthalmologist once advised his students "never write on nystagmus, it will lead you nowhere."[7]

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One rare type of nystagmus, rebound nystagmus, has been described only in patients with cerebellar disease.[8-10] To test for this nystagmus, the patient first looks in one direction (say to the right). In patients with a positive response, a brisk nystagmus with its fast component to the right appears. If the patient continues looking in this direction for about 20 seconds, the nystagmus fatigues and disappears (sometimes even reversing direction). The patient then returns his or her eyes to the primary position (i.e., straight ahead) and nystagmus to the left, not present initially, appears, although it fatigues over time. In these patients the direction of the nystagmus in primary gaze can be reversed at will, depending on whether the patient looks first to the left or the right.[8]

B. NYSTAGMUS AND NONCEREBELLAR DISORDERS

Other useful patterns of nystagmus (not features of cerebellar disease) are optokinetic nystagmus (see Chapter 58), the nystagmus of internuclear ophthalmoplegia (see Chapter 59), and the nystagmus of vestibular disease (see Chapter 68).

3. EFFECT OF RETINAL FIXATION

Retinal fixation means that the patient is focusing his or her eyes on an object. Spontaneous nystagmus that diminishes during retinal fixation argues that the responsible lesion is located in the peripheral vestibular system; nystagmus that increases or remains unchanged during fixation argues that the lesion is in the central nervous system (i.e., brainstem or cerebellum). Neuro-ophthalmologists usually use electronystagmography to detect the effects of fixation (by comparing eye movements with eyes open with those with eyes closed), but general clinicians can accomplish the same during direct ophthalmoscopy: in a dimly lit room, the clinician examines the optic disc of one eye and, as the patient fixes the opposite eye on a distant target, compares its movements with those when the patient's opposite eye is covered. If rhythmic movements of the optic disc first appear or worsen when the fixating eye is occluded, a peripheral vestibular disturbance is likely.[11] A simpler version of this test using just a penlight without ophthalmoscopy has been proposed.[12]

C. HYPOTONIA (SEE CHAPTER 61)

The limbs of patients with cerebellar disease offer no resistance to passive displacement, sometimes resembling (in the words of Gordon Holmes) the "muscles of a person deeply under an anesthetic, or of a corpse recently dead."[1] Holding the forearms vertically causes the wrist to bend to an angle much more acute than normal. Displacing the patient's outstretched arm downward causes abnormally wide and prolonged up-and-down oscillations, even when the patient is asked to resist such movements. Striking the patellar tendon causes pendular knee jerks, traditionally defined as three or more swings,[13] although, as already stated in Chapter 61, this threshold will have to be revised upward because many normal persons also demonstrate three or more swings.[14]

D. DYSARTHRIA

The speech of patients with cerebellar disease is slow, slurred, and irregular in volume and rhythm, findings that are collectively referred to as **dysarthria**. In contrast to patients with aphasia, however, patients with dysarthria can name objects, repeat words, comprehend language, and speak sentences with words whose order makes sense.

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TABLE 65.1 Unilateral Cerebellar Lesions[13,15*]

Physical Finding†	Frequency (%)‡
Ataxia	
Gait ataxia	80-93
Limb ataxia	
Dysmetria	71-86
Intention tremor	29
Dysdiadochokinesia	47-69
Nystagmus	54-84
Hypotonia	76
Pendular knee jerks	37
Dysarthria	10-25

**Diagnostic standard*: clinical imaging, surgical findings, or postmortem examination.
†Definition of findings: see the text.
‡Results are overall mean frequency or, if statistically heterogeneous, the range of values.
Data from 444 patients.

III. CLINICAL SIGNIFICANCE

A. INDIVIDUAL FINDINGS

1. ATAXIA

Ataxia of gait is the most common finding in all cerebellar syndromes (Table 65.1); therefore, examination of the gait should be part of the evaluation of any patient with suspected cerebellar disease. Many patients with cerebellar disease have difficulty walking despite the absence of all other findings of limb ataxia.

Simple measurements of the patient's dysdiadochokinesia—such as how quickly and accurately the patient can alternately tap two buttons spaced about 12 inches apart†—are accurate measures of ataxia that correlate well with other measures of disability.[16]

2. NYSTAGMUS

Seventy-five percent of cerebellar nystagmus is a conjugate horizontal jerk nystagmus that appears on lateral gaze (15% is a rotatory nystagmus and 10% a vertical nystagmus). Nonetheless a horizontal jerk nystagmus is not specific for cerebellar disease; it also occurs in peripheral vestibular disease and other central nervous system disorders. The direction of the jerk nystagmus has less localizing value than tests of ataxia. (See the section titled "Cerebellar Hemisphere Syndrome.")

The clinical utility of rebound nystagmus is limited because it is a late finding, and all patients described with the finding have had many other obvious cerebellar signs.[8,9]

3. DYSARTHRIA

Dysarthria, the least common of the fundamental cerebellar signs (see Table 65.1), appears more often with lesions of the left cerebellar hemisphere than with those of the right hemisphere.[17]

†Ninety percent of normal persons can accomplish at least 32 taps within 15 seconds, whereas 90% of patients with cerebellar ataxia cannot.[16]

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B. CEREBELLAR SYNDROMES

Most patients with cerebellar disease present with difficulty walking or headache or both.[13,15] In adults there are four common cerebellar syndromes, each of which is characterized by a different distribution of the principal cerebellar signs.

1. CEREBELLAR HEMISPHERE SYNDROME

A. CEREBELLAR FINDINGS

Table 65.1 presents the physical findings of 444 patients with focal lesions (mostly tumors) confined to one hemisphere.[13,15] According to traditional teachings, cerebellar signs appear on the side of the body *ipsilateral* to the lesion. This teaching proved generally correct in the patients of Table 65.1, in whom signs of limb ataxia (i.e., dysmetria, intention tremor, dysdiadochokinesia) were unilateral 85% of the time and, if unilateral, were on the side ipsilateral to the lesion 80% to 90% of the time. These patients also had more hypotonia on the side of the lesion and tended to fall toward the side of the lesion when walking.

Nystagmus has less localizing value. When present, nystagmus is unilateral in only 65% of patients; in these patients the direction of nystagmus points to the side of the lesion only 70% of the time.

B. ASSOCIATED FINDINGS

Despite having a lesion confined to the cerebellum, patients with structural cerebellar lesions may also have (1) cranial nerve findings (10% to 20% of patients; usually of cranial nerves V, VI, VII, or VIII ipsilateral to the side of the lesion 75% of the time);[13,15] (2) altered mental status (38% of patients, from compression of the brainstem or complicating hydrocephalus); (3) upper motor neuron signs such as hyperactive reflexes and the Babinski sign (28% of patients); and (4) papilledema (68% of patients).

In contrast, severe weakness and sensory disturbance are both uncommon, affecting only 4% of such patients.

2. ANTERIOR CEREBELLAR DEGENERATION (ROSTRAL VERMIS SYNDROME)[18]

In contrast to the cerebellar hemisphere syndrome, these patients have ataxia of gait (100%) and of both legs (88%) with relative sparing of the arms (only 16% of patients). Nystagmus and dysarthria are also much less frequent (9%, for both findings). This syndrome most often results from chronic alcohol ingestion.

3. PANCEREBELLAR SYNDROME

This syndrome causes the same signs as listed in Table 65.1, but instead of being on one side of the body, the cerebellar signs are symmetric. Causes include drug intoxication (e.g., phenytoin), inherited disorders, and paraneoplastic syndromes.

4. CEREBELLAR INFARCTION

The physical signs of cerebellar infarction resemble those of the cerebellar hemisphere syndrome described above, with three exceptions. In infarction, (1) all signs appear *abruptly*, (2) dysarthria is more prominent (44% of patients), and (3) weakness occurs more often (22% of patients have hemiparesis and 24% have tetraparesis).[19-22] The three main arteries supplying the cerebellum are the superior cerebellar artery, anterior inferior cerebellar artery, and posterior inferior cerebellar artery.[23] An associated lateral medullary syndrome (see Table 62.2 in Chapter 62) suggests an infarct in the distribution of the posterior inferior cerebellar artery.[21,24]

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The **acute vestibular syndrome**—the abrupt onset of sustained vertigo, nausea and vomiting, and imbalance—raises the possibility of cerebellar infarction as well as peripheral vestibular disease. This subject is fully discussed in Chapter 68.

The references for this chapter can be found on www.expertconsult.com.

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